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Juvenile Idiopathic Arthritis and OT role

Cameron Van Oort BSc, MSc, MScOT, CHT

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Non-financial: Cameron Van Oort has no relevant non-financial relationships to disclose.

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Learning Outcomes

After this course, participants will be able to:

- Discuss the disease process of JIA in relative detail.
- Recognize the various impacts and sequelae of JIA.
- Identify interventions for children with JIA.

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Synovial Joint Structure

- Synovial joints
  - Two or more bone ends meet
  - Articular cartilage
    - Fibro & hyaline cartilage
  - Articular capsule
    - Subintima & intima
  - Joint cavity
    - Synovial fluid
  - Ligaments
    - Intracapsular or Extracapsular

(Armiento et al, 2019)
Arthritis

- *Arthro* = joint  *itis* = inflammation
- Osteoarthritis
  - Positive feedback loop
- Rheumatoid arthritis
  - Autoimmune
- Inflammation leads to:
  - Joint laxity
  - Obstructive bone growth

Juvenile Idiopathic Arthritis

- A heterogeneous group of diseases diagnosed in children <16 years of age
- Autoimmune disease
- 7 subtypes
  1. Systemic
  2. Oligoarticular/Pauciarticular
  3. Polyarticular rheumatoid factor positive
  4. Polyarticular rheumatoid factor negative
  5. Enthesitis-related
  6. Psoriatic
  7. Undifferentiated

(Barut K et al, 2017)
Common Symptoms

- Arthritis
- Arthralgia
- Uveitis
- Limping
- Fever
- Skin rash
- Abdominal pain

(Aoust et al, 2017)

Etiology

- Idiopathic. However…

- Genetic factors
  - > prevalence in siblings and twins
  - HLA and various other gene complexes

- Environmental factors
  - Bacterial infections
  - Viral infections

(Rigante et al, 2015)
Diagnosis

- <16 years of age
- >6 weeks of symptoms
- Excludes other conditions (differential diagnoses)

- No single laboratory test to confirm diagnosis
  - CRP & ESR
  - X-ray, ultrasound, MRI

(Epidemiology)

Average age of onset is 4.8 years of age
Incidence rate of 5.7/100,000 for boys vs. 10.0/100,000 for girls
Prevalence rate of 11.0/100,000 for boys vs. 19.4/100,000 for girls

Differences also:
- Geographically (vitamin D levels)
- Racially
- Subtypes
- Gender

(Giancane et al, 2016; Harold et al, 2013; Kuntze et al, 2018; Thierry et al, 2014)
Epidemiology

- Oligoarticular is most common subtype
- Most common joints involved
  1. Knees
  2. Hands
  3. Feet
- Continues in adulthood in ~30% of patients

Physical Impact

- Swelling
- Reduced physical activity
- Reduced strength
- Reduced ROM (e.g., contractures)
- Reduced function
- Reduced bone mineral density
- Muscle atrophy

(Borchers et al, 2006; Harrold et al, 2013; Giancane et al, 2016; Miedany et al, 2019)

(Lindehammer & Lindvall, 2004; Cavallo et al, 2014)
Psychosocial Impact

- Chronic pain
  - Increased pain sensitivity
  - Hyperalgesia
  - Allodynia
- Sleep disturbances
- Reduced leisure activity
- Negative impacts on schooling
- Reduced quality of life


Psychosocial Impact

- Internalizing behaviors
  - Emotional difficulties/Reduced mood
  - Low self-esteem
  - Distorted self-image
  - Anxiety
  - Depression
- Externalizing behaviors
  - Aggressive behaviors
  - Rule-breaking behaviors

Margetic et al, 2005; van der Meer et al, 2007; Memari et al, 2016
Remission

- #1 goal with treatment is remission. Otherwise...
  - Erosion of bone and cartilage
  - Growth disturbances
  - Joint fusion
  - Malalignment

- Remission types
  - Clinically inactive disease
  - Remission on medication
  - Remission off medication

(Muller et al; 2015; Shoop-Worrall et al, 2017)

Positive correlation between disease duration and remission frequency

(Shoop-Worrall et al, 2017)
THE OT ROLE

ASSESSMENT
The key to developing effective interventions
General Assessments

- COPM
  - What are the child and/or caregivers hoping for?
- F-Words in Childhood Disability
- PedsQL
  - Physical, emotional, social and school functioning
  - 2-18 years of age
  - License fee

(Sturgess et al, 2002; Varni et al, 2002; CanChild, 2020)

Disease-Specific Assessments

- Childhood Health Assessment Questionnaire (CHAQ)
  - Disability index, Discomfort index and Health status
  - 1-19 years of age
  - Free for research purposes; no materials needed
- POSNA Pediatric Musculoskeletal Functional Health Questionnaire
  - Disability, Pain and Health
  - 2-18 years of age
  - Free; no materials needed
Disease-Specific Assessments

- Juvenile Arthritis Functional Assessment Report (JAFAR)
  - Ability to perform physical tasks
  - 7-18 years of age
  - Free; No equipment needed

- Juvenile Arthritis Quality of Life Questionnaire (JAQQ)
  - Motor skills, psychosocial function, general symptoms and pain
  - 2-18 years
  - Free; No equipment required

- Juvenile Arthritis Functional Assessment Scale (JAFAS)
  - Assesses ADLs considered important in children with JIA
  - 7-16 years of age
  - Free; Minor cost for materials

- Juvenile Arthritis Functional Status Index (JASI)
  - Functional ability assessment
  - 8-17 years of age
  - $25 USD; no materials needed

Physical Assessment

- Number and location of joints involved
- ROM (goniometry)
- Strength (MMT, dynamometry)
- Swelling (circumferential, figure-of-eight, volumetric)

(Petersen et al, 1999)

Psychosocial Assessment

- Pain (VAS, NRS, MPQ, APPT)
- Conners Early Childhood (Conners EC)
  - Ages 2-6 years (fee for use)
- Child Behavioral Checklist (CBCL)
  - Ages 6-18 years (fee for use)

(Jacob et al, 2014; Memari et al, 2016)
Interventions
A Collaborative Process

Pharmaceutical Interventions

- NSAIDs
  - Ibuprofen, Naproxen, Indomethacin
- Intra-articular corticosteroid injections
- Prednisone (oral corticosteroid)
- DMARDs
  - Methotrexate, Leflunomide
- Biologics
  - Etanercept, Infliximab, Adalimumab, Golimumab

(Giancane et al, 2016)
Exercise

- Improves aerobic fitness
- Improves strength and function
- Improves self-efficacy
- Improve QoL
- Reduces disease activity

- Low-impact for 30-50 minutes, 2-3x/wk. with a focus on strength, balance and flexibility

(Philpott et al, 2010; Kuntze et al, 2019; Klepper et al, 2019)

Orthotic Intervention

- Medication has greatly improved in last decade, reducing need for orthosis.
- Most splinting research relates to the wrist and knee, with conflicting evidence
  - Splinting after cortisone injection
- Serial casting for PIP joint contractures
- Children may be resistant to wearing splints due to social stigma

(Helders et al, 2002; Schroder et al, 2002; Wallen & Gillies, 2008; Dunbar et al, 2016; Ugurlu & Ozdogen, 2016; Naz et al, 2018)
Adaptive Aids

- The goal with adaptive aids to reduce pressure on the affected joints and ideally shift the load to larger or more joints
  - Extended comb handles/long handled bathing sponge
  - Adaptive eating utensils (e.g., thicker spoon handles)
  - Shoe horn
  - Velcro on clothing and/or shoes.
  - Elevated toilet seat
  - Wheelchairs
  - Writing aids
  - Dycem

(Cakmak & Bolukbas, 2005)

Psychological Intervention

- Evidence for psychological interventions in JIA is conflicting. However,
  - Better outcomes for children with low quality of life compared to high quality of life
  - Better outcomes with peer-support groups relative to individual based therapies

- Types of interventions researched:
  - Mind-body interventions
  - Cognitive-behavioral interventions
  - Peer-support interventions

(Fuchs et al, 2013; Cohen et al, 2017; Waite-Jones and Swallow, 2018)
Liam

- 8-year-old boy
- Significant fever 18 months ago that persisted and began getting significant pain along with significant fatigue and reduced function.
- Diagnosed 1 year ago with systemic JIA
- Currently on biweekly injections of Enbrel and oral Naproxen 2x/day
Assessment

- F-Words of Child Disability
- JAFAR
- ROM measurements
- Pain
Table 4. The Pediatric Arthritis Functional Assessment Report for Children (PAFAR-C)∗

<table>
<thead>
<tr>
<th>Task</th>
<th>All the time</th>
<th>Sometimes</th>
<th>Almost never</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Take shirt off hanger</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>2. Button snaps</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>3. Pull on sweater over head</td>
<td></td>
<td></td>
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<tr>
<td>4. Turn on water faucet</td>
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<tr>
<td>5. Lunch in helmet</td>
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<tr>
<td>6. Dry brush with towel</td>
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<tr>
<td>7. Brush face with washcloth</td>
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<td></td>
<td></td>
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<tr>
<td>8. Tie shoes</td>
<td></td>
<td></td>
<td></td>
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<tr>
<td>9. Pull on socks</td>
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<td></td>
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<tr>
<td>10. Brush teeth</td>
<td></td>
<td></td>
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</tr>
<tr>
<td>11. Stand up from chair without using arms</td>
<td></td>
<td></td>
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</tr>
<tr>
<td>12. Go up steps</td>
<td></td>
<td></td>
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</tr>
<tr>
<td>13. Cut hand with both and fork</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>14. Lk or easy glass of water</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>15. Women pericentrum escorted trunk</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>16. Walk 5 feet without help</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>17. Walk up 5 stairs</td>
<td></td>
<td></td>
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<tr>
<td>18. Stand up on stairs</td>
<td></td>
<td></td>
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</tr>
<tr>
<td>19. Reach above head</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>20. Get out of bed</td>
<td></td>
<td></td>
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</tr>
<tr>
<td>21. Pick up something from floor from standing position</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>22. Push open door after turning knob</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>23. Turn head and look over shoulder</td>
<td></td>
<td></td>
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</tr>
</tbody>
</table>

*Recommend scores if the all the time, 1 for sometimes, and 3 for almost never. On the PAFAR the Patient's parent should be asked. If they do not work, have attention was directed with the response alternative, move and be interviewed in follow-up. Howe et al (1991 with permission from Arthritis Care & Research)

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ROM

- Joints with potential limit or pain
Pain

- Adolescent Pediatric Pain Tool
- Location
- Intensity
- Quality
- Temporal pattern

Interventions

Basing the intervention around the goals:

1. School work
   - Speech-to-text
   - Scheduling breaks/More time for assignments

2. Traveling with family
   - Bathing in mornings to warm up/loosen joints
   - Adequate time between activities
   - Wheelchair
Interventions

3. Gain strength
   • Education on the benefit of an exercise program targeting strength, flexibility and balance

4. Video games with friends
   • Adaptive controllers
   • Varying styles of games
     • Gross motor games (Nintendo Wii)
     • Fine motor games (Xbox, Playstation)

5. New friends with JIA
   • Organize a peer support group
   • Education on various camps
     • https://campcambria.org/ (Minnesota & Ontario)

6. Riding a bike
   • Continue with strengthening
   • Educating parents that activity will not harm child
   • Adaptive aids if necessary (e.g., electric bike, higher handle bars, thicker handle bars, etc.)
References

- Aoust L, Rossi-Semerano L, Koné-Paut I, Dusser P. 2017. Time to diagnosis in juvenile idiopathic arthritis: a french perspective. Orphanet J Rare Dis, 12(43) 1-5.
References

References


Questions?

- Email: cvanoor@uwo.ca